September 12, 2003

TO: ALL COUNTY CALIFORNIA CHILDREN’S SERVICES (CCS) PROGRAM ADMINISTRATORS, MEDICAL CONSULTANTS, CHILDREN’S MEDICAL SERVICES (CMS) BRANCH AND REGIONAL OFFICE STAFF, AND MEDICAL DIRECTORS OF CCS-APPROVED SPECIAL CARE CENTERS

SUBJECT: MEDICAL ELIGIBILITY NEPHROTIC SYNDROME

The purpose of this numbered letter is to provide local county CCS programs and CMS Branch Regional Offices with guidance for determining medical eligibility for children diagnosed with Nephrotic Syndrome.

I. Background

CCS medical eligibility regulations, Title 22, California Code of Regulations, Division 2, Subdivision 7, Section 41856 (b), filed July 7, 2000, state that “CCS applicants with at least one of the following conditions shall be medically eligible for participation in the CCS Program: Chronic Glomerulonephritis, Chronic Nephrosis, or Chronic Nephrotic Syndrome”.

Nephrotic Syndrome is a clinical condition characterized by massive urinary loss of protein (primarily albumin), which leads to hypoproteinemia and edema. Nephrotic Syndrome is categorized as either primary Nephrotic Syndrome, also known as Nephrosis, and secondary Nephrotic Syndrome. In primary Nephrotic Syndrome, there is no clear underlying cause. Minimal-change Nephrotic Syndrome accounts for approximately 80 percent of cases of primary Nephrotic Syndrome. Secondary Nephrotic Syndrome is associated with an underlying disease, such as anaphylactoid purpura, systemic lupus erythematosus, diabetes mellitus, sickle cell disease, and syphilis.
Most children with primary Nephrotic Syndrome will respond to corticosteroids within the first two to eight weeks of treatment. Those who do not initially respond may be non-responsive to steroids, steroid dependent, or may have frequent or infrequent relapses. Approximately three-quarters of those who respond to initial treatment will have at least one relapse and close to 50 percent will have multiple relapses.

It is important that appropriate care be provided from the time of diagnosis for all children with Nephrotic Syndrome. At the onset of the disease, it is impossible to determine who will respond to initial treatment and never have a relapse and who will progress to have chronic disease that is non-responsive to treatment or relapses.

II. Policy and Policy Guidelines

1. All children and adolescents diagnosed with Nephrotic Syndrome who meet CCS Program residential and financial eligibility shall be eligible for the CCS Program.

2. All CCS-eligible children and adolescents diagnosed with primary Nephrotic Syndrome shall be authorized to receive care from a CCS-paneled Nephrologist at a CCS-approved Renal Special Care Center at the time of initial diagnosis.

   Authorizations to a CCS-paneled Nephrologist in the local community may also be issued with documentation of care coordination between the local physician and the CCS-approved Renal Special Care Center.

3. Children and adolescents diagnosed with secondary Nephrotic Syndrome and who are already authorized to receive care at a CCS-approved Special Care Center for their primary medical condition:

   a) Shall have care for Nephrotic Syndrome provided by the authorized Special Care Center team and consultant staff if so designated by the Special Care Center Director.

   b) Shall be authorized at a CCS-approved Renal Special Care Center for care related to Nephrotic Syndrome if the medical director of the CCS-approved Special Care Center authorized to provide care for the primary medical condition determines that this is in the best interest of the child or adolescent. If an authorization is issued to a second Special Care Center, a clear plan for coordination between the core team members of the two centers must be reviewed and approved by the county or Regional Office Medical Consultant or designee.
4. Children and adolescents diagnosed with secondary Nephrotic Syndrome and whose primary CCS-eligible medical condition is newly diagnosed or not cared for in a CCS-approved Special Care Center, care for the Nephrotic Syndrome shall be authorized to a CCS approved Renal Special Care Center.

5. Authorizations to a CCS-approved Renal Special Care Center shall be issued for a period of one year or until the date of the child’s next program eligibility determination.

If you have any questions regarding this policy, please contact your Regional Office Medical Consultant.

Original Signed by Maridee Gregory, M.D.

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Children’s Medical Services Branch